Idiopathic Intracranial Hypertension: Diagnostic Accuracy of the Transverse Dural Venous Sinus Attenuation on CT Scans

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SUMMARY – *Idiopathic intracranial hypertension (IIH) is a clinical disorder of unknown etiology. It may exhibit several non-specific imaging findings. We observed that patients with intracranial hypertension demonstrate intracranial venous sinus attenuation with changes in the contour and caliber of the distal transverse sinus. This can be seen on routine non-contrast sagittal reformatted CT images of the brain. We have termed this the venous attenuation sign (VAS). This study evaluated the VAS as a marker for IIH assessing the transverse sinuses on sagittal reformatted non-contrast CT for the presence of a VAS in 25 patients with IIH and 24 control patients. Scans were independently assessed in a blinded fashion by three readers. The readers identified the VAS in 96% of patients with IIH; 83.3% of the control patients were negative for VAS. Our study supports the VAS as an additional imaging marker which may be incorporated into the evaluation of patients suspected to have this condition.*

Introduction

Idiopathic intracranial hypertension (IIH), also known as pseudotumor cerebri, is a syndrome characterized by increased intracranial pressure of unknown cause [1-5]. It predominantly affects overweight women of childbearing age [2,3]. The yearly incidence of IIH is approximately 1 per 100000 [2].

Patients usually present with headaches, nausea, vomiting, visual field disturbances and classically papilledema on physical examination [3,7]. Diagnosis is based on the modified Dandy criteria, which include raised lumbar puncture (LP) opening pressure in the absence of an intracranial mass lesion or ventricular dilatation, normal spinal fluid composition, normal neurological examination except for papilledema and occasional VI or VII nerve palsies, and normal level of consciousness [6].

Proposed mechanisms of pathogenesis for IIH revolve around cerebrospinal fluid (CSF) physiology [8-12]. Recently, vascular causes have also been proposed to contribute to the underlying pathology, with emphasis on arterial inflow and venous outflow abnormalities [13-16].

Early diagnosis and treatment are important because they can preserve vision. However, this can be difficult due to the variety of clinical presentations. Additional diagnostic tools, such as neuroimaging, may be helpful in assisting diagnosis [17-19]. Traditionally, the role of imaging in the evaluation of IIH has been to exclude secondary causes of increased intracranial pressure (ICP) and papilledema [7,20]. Several radiological findings have been described in the literature that may aid in establishing the diagnosis such as: flattening of the posterior aspect of the globe, increased perioptic nerve sheath dimensions, and the presence of an empty sella [19,21-28]. Previous reports suggest that most of these findings aid in the diagnosis of IIH, but lack sufficient specificity [29].

On review of the CT imaging of patients presenting to our institution with IIH, we have identified a possible novel imaging sign for the

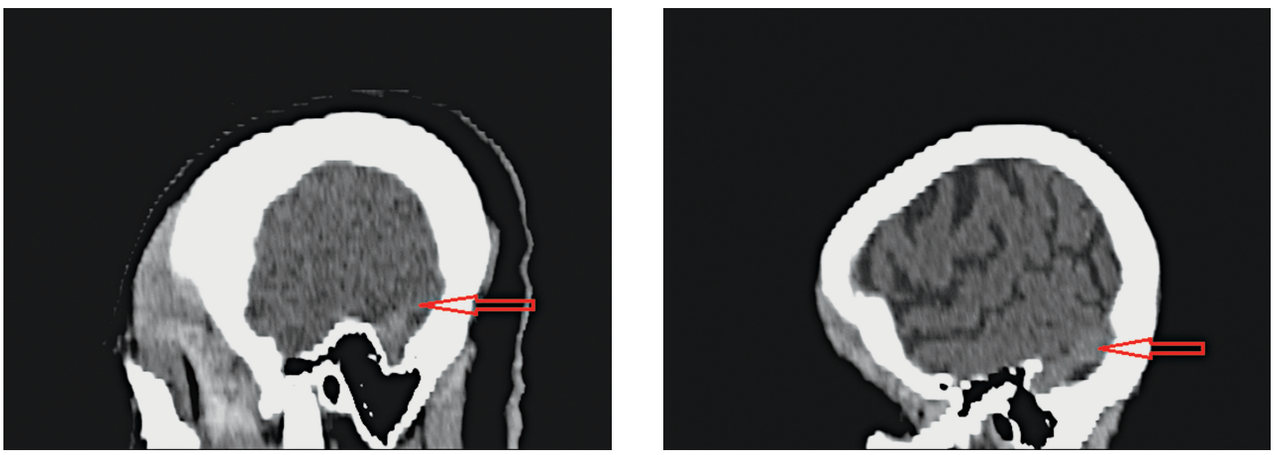

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Figure 1 Non-contrast CT scan sagittal reformatted images at the level of distal transverse sinus. A) A positive VAS is demonstrated in a patient known to have IIH proved by lumbar puncture. Note the sinus is thin with straight upper and lower borders (red arrow). B) Normal sinus appearance in a control patient with normal CSF pressure on lumbar puncture. The sinus has an appreciable luminal diameter with concave borders (red arrow).

diagnosis of IIH. The sign, referred to in this paper as the venous attenuation sign (VAS) can be evaluated on routine non-enhanced reformatted sagittal CT images of the brain, where the sinus is identified by its normal relative hyperdensity to adjacent brain parenchyma. This is best seen at the level of the lateral aspect of at least one transverse sinus (TS) (the dominant sinus) (Figures 1 and 2).

The objectives of this study were: a) to evaluate VAS as a marker for IIH and b) to analyze the frequency of traditionally reported radiological manifestations of IIH (i.e. empty sella, optic nerve (ON) sheath distension, ON vertical tortuosity, posterior globe flattening and intraocular protrusion of ON head) in our patient population.

Materials and Methods

This study was approved by the institutional review board.

Patients with IIH

We reviewed the clinical and imaging records of sequential patients who underwent LP for evaluation of IIH at our institution (over a ten-month period) and had a non-contrast CT within three months of LP.

The reason provided for the examinations was headache. The diagnosis of IIH was established by clinical signs of papilledema with non-focal neurologic examination, and CSF studies revealing normal composition and lumbar puncture opening pressure greater than 20 cm H_2O. Patients with a history of trauma, congenital cranial malformation, subarachnoid hemorrhage, intracranial surgery unrelated to IIH, and those with secondary causes of increased ICP on imaging, such as hydrocephalus, intracranial mass, cerebral venous thrombosis or chronic meningitis, were excluded.

The study population consisted of 25 patients with IIH (22 women and 3 men) with an average age of 33.2 years (range: 10-81 years).

Control subjects

An age and sex matched control group was selected from our PACS station. They were headache patients with normal lumbar puncture opening pressure and normal fluid composition and with normal non-contrast CT imaging. Control patients were on average 31.3 years (range: 18-60 years). The CSF opening pressure ranged from 7-20 cm H_2O.

Imaging, processing and review

CT imaging

Non-contrast CT imaging was obtained in all patients (IIH group and control group) using multislice CT scanners. Our examinations were performed using PHILIPS BRILLIANCE 64 and

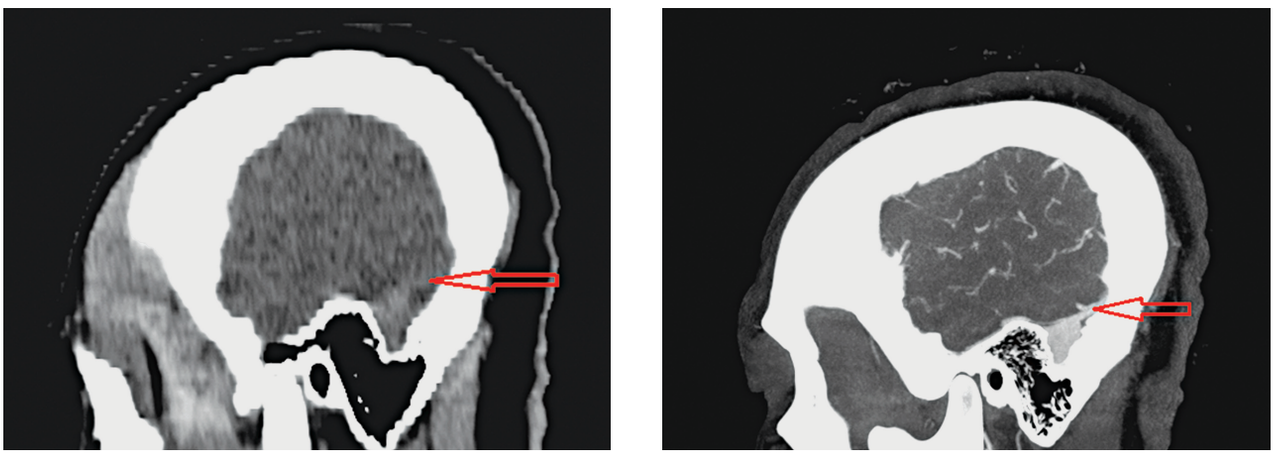

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Figure 2 Images of patient with IHH. A) Non-contrast CT sagittal reformatted image showing positive VAS. B) This is proved on the post-contrast CT study (red arrows).

16 slice CT scanners. Images were transferred to a PACS viewing workstation (GE Advantage windows 4.2) where they were reformatted in the sagittal plane. Image scan parameters were: 16 slice scanner: standard resolution, 120 kVp, 440 mAs, 0.5 second rotation time, 16 × 0.75 mm collimation, 0.563 pitch, filter: UB, 3 mm slice thickness, 3 mm increments, window: 35/70, 14 second scan time, 67 mGy $CTDI_{vol}$; 64 slice scanner: standard resolution, 120 kVp, 550 mAs, 1.5 second rotation time, 16 × 0.625 mm collimation, 0.563 pitch, filter: UB, 3 mm slice thickness, 3 mm increments, window: 35/70, 4.8 second scan time, 28.3 mGy $CTDI_{vol}$. Contrast CT study was performed in two patients where the VAS was also demonstrated (Figure 2B).

The studies were interpreted by three neuroradiologists (RM, AD and OM). Each reader was independently trained for several minutes on the use of the VAS using examples of control and confirmed IIH in cases other than the study population. To standardize the interpretation, the larger (dominant) of the two transverse sinuses was used. The VAS was deemed positive if the dominant sinus appeared thin and thread-like with straight borders. It was deemed negative if appreciable sinus lumen was identified on the dominant side showing concave borders.

The readers were asked to tabulate whether the VAS was present or absent for each image set. The readers were given 49 randomly ordered studies. They were unaware of the number of patients with IIHs versus normal controls. The readers were blinded to all patient information. Each reader independently reviewed the 49 sets of images. The results of the three readers were grouped for each patient and a consensus response was derived based on the majority opinion (two of three).

MRI imaging

Twenty-two patients with IIH had MRI examination of the brain within days to weeks after the non-contrast CT. The MRI examinations were performed on a superconducting 1.5 T unit with a head coil. Our examinations were done using a 1.5T GE Signa scanner. The foreign MRI examinations were transferred to our PACS station. All subjects were scanned using a standard protocol for headaches with a section thickness 4.0 mm and a gap of 30%, including axial T1, T2WI, axial FLAIR, diffusion-weighted, and sagittal T1 sequences.

Post-contrast images were obtained in 14 cases in the axial, sagittal, and coronal planes. Each set of CT and MRI scans were assessed by two neuroradiologists for the presence of: 1) flattening of the posterior sclera relative to the curvature of the globe, 2) distension of the perioptic subarachnoid space, 3) intraocular protrusion of the prelaminar ON into the vitreous cavities, 4) tortuosity of the orbital ON, and 5) empty sella.

Partially empty sella was considered present when the majority of the sella turcica was filled with CSF with the presence of a thinned

pituitary gland at the base of the sella. A completely empty sella was diagnosed if the pituitary fossa was completely filled by CSF with no visible pituitary gland.

Results

Performance of the VAS as a test

VAS sensitivity was 96% (80-99%), 95% CI. The three readers correctly identified the IIHs with a VAS in 24 of 25 subjects (unanimously in 17 subjects and by majority [two of three] in seven subjects). One subject was only correctly identified by one of the three readers. Specificity of the sign was 83% (64-93 %), 95% CI. The three readers correctly identified the control patients with a negative VAS in 20 of 24 subjects (unanimously in 14 subjects and by majority in six subjects). Four control patients were interpreted to have a positive VAS by majority [two of three readers]. The positive predictive value was 86% (68-94%), 95% CI. The negative predictive value was 95% (77-99%), 95% CI. The positive likelihood ratio was 5.7 (2.3-14), 95% CI, while the negative likelihood ratio was 0.05 (0.007-0.3), 95% CI. The accuracy of the VAS was 0.9.

Cross sectional MRI/ CT findings in IIH

Previously reported IIH imaging findings were assessed for the 25 IIH patients; findings are summarized in Table 1. Empty sella was found in 17 subjects (68%) [seven empty sella + ten partially empty sella].

Table 1 Presence of previously reported [47] imaging signs of IIH in among patients diagnosed with IIH.

Sign	*No of cases*
Venous attenuation sign	24/25(96%)
Empty/partially empty sella	17 (68%)
Flat posterior sclera	16 (64%)
Dilated optic nerve sheath	16 (64%)
Tortuous optic nerves	6 (24%)
Intraocular protrusion of optic nerve head	4(16%)
Combination of 4 signs	5 (20%)
Combination of 3 signs	10(40%)
Combination of two signs	2 (8%)
Only one sign (empty sella)	1 (4%)

Discussion

Our study describes a new sign, the venous attenuation sign (VAS), which can be helpful in the diagnosis of IIH. If this sign is present, suspicion can be raised and lumbar puncture could be performed. We found that the VAS has a high sensitivity (96%) and specificity (83.3%) for detection of IIH with a high positive predictive value (85.7%). To our knowledge, VAS in IIH has not been described in the literature.

As shown in the present study, VAS can be applied to the non-enhanced CT scan to provide insight into the status of intracranial pressure early in the diagnostic workup.

There are a few CT signs like empty sella sign which are suggestive of IIH. In our study, 68% of the IIH group had complete or partially empty sella. The incidence of empty sella in cases of IIH varies widely (10-94%), probably related to discrepancies in definition [26]. Although empty sella is associated with IIH, it is also present in 5-6% of normal individuals [26,45]. Therefore, it should probably be considered a normal variant in the vast majority of asymptomatic patients. The ON and globe findings are significant in that they probably better predict papilledema [46]. In a review of the MRI findings of 30 patients, Agid et al. [47] found that four of the traditional imaging findings were significantly associated with IIH (empty sella, posterior globe flattening, ON sheath distention, ON tortuosity). However, they concluded that only posterior globe flattening had a high enough specificity to be of clinical use. Lim et al. studied similar MRI findings in 23 children, finding that a combination of three or more findings of globe flattening, empty sella, protrusion of the ON head, and tortuosity of the ON would increase specificity to 95% for the diagnosis of IIH [48]. However, the combination of three or more of these findings was only present in 60 % of our IIH group.

Bilateral narrowing of the transverse sinus (TS) without associated thrombosis is a common finding among patients with IIH [31-35]. One theory is that the stenosis causes IIH. Angiographic catheters passed into the intracranial venous sinuses through the jugular veins measured elevated intrasinus pressures in many IIH patients [36,40]. Sometimes these high pressures appeared to be secondary to central venous hypertension, but more often they seemed to be the result of stenotic lesions in the lateral sinuses obstructing cerebral venous outflow. Further credence is lent to this idea

by the fact that stent angioplasty can help control intracranial hypertension (IH) in some patients [33,34,42-44].

However, lowering the intracranial pressure can result in normalization of venous morphology, suggesting TS stenosis might be induced by IH itself (secondary cranial venous outflow obstruction (CVOO)) [36,37]. This has led one group to propose intracranial venous hypertension as the final common pathway in the etiology of IIH [36]. Riedel et al. have shown that CVOO is not a specific feature of patients with IIH but a universal phenomenon in most patients with IH irrespective of the underlying cause. A widespread involvement of the venous sinuses was associated with very high CSF pressure. They hypothesized that IH compresses the dural sleeve of the venous sinuses [38]. Regardless of cause or effect, patients with IIH are likely to have raised venous pressures and stenotic lesions in their lateral venous sinuses. A study using a technique of contrast-enhanced magnetic resonance venography (MRV) found that narrowing of the distal transverse sinuses was strongly associated with IIH. It was also shown that application of a simple grading scheme for the degree of sinus narrowing provided a highly sensitive (93%) and specific (93%) test for identifying patients with IIH [41]. Higgins et al. concluded that MRV in patients with IIH commonly presents a pattern of bilateral lateral sinus defects rarely seen in asymptomatic controls [30].

Our study has several limitations. It is retrospective and has relatively few patients. A correlation with venographic studies is lacking. Future studies can address these issues. Additionally, because this is the first time this sign is described it is uncertain what predictive power it would have in general use. Finally, the authors' training set was relatively small, and it is possible that accuracy would rise if VAS were assessed routinely.

In conclusion, the venous attenuation CT sign could be used to suggest IIH and prompt further work-up in the appropriate clinical setting.

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